Modern Concepts of Cardiovascular Disease

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THE PROGNOSIS IN RHEUMATIC HEART DISEASE

TO DETERMINE prognosis in rheumatic heart disease is difficult because so little is known of the natural course of the disease. This is so chiefly because the disease is characterized, as a rule, by a slow, gradual progression towards a fatal termination many years after the initial infection. It is almost impossible, therefore, to observe the entire course of the disease in large numbers of individuals. Furthermore, onset, though frequently dramatic and thereby definitely recognized, is often insidious and hard to determine. Because the disease is so common in children it is generally believed by pediatricians that it always begins in childhood. When it is seen in adults, they believe that an insidious infection and slow progression of the disease in early life was overlooked or forgotten in later years, and that this leads to an erroneous impression that a subsequent recurrent manifestation represents the initial affection. Increasing evidence has been accumulating, on the other hand, based on clinical as well as post mortem examinations, that initial attacks of rheumatic fever are experienced in adult life.

When a patient is first seen, in order to forecast his future course, one should determine how far the disease has progressed. To do this the examination of the patient should be supplemented by a careful history which should elicit if possible the date of initial rheumatic infection, the date of discovery of heart disease, when symptoms of diminishing cardiac reserve were first noted and when the first heart failure occurred. In making such predictions, however, it must be remembered that they apply to groups, not to individuals. Prediction of the course of disease in any individual can be made only in terms of probability.

It has been shown in a series of adult patients recently reported that the average duration of life of a rheumatic cardiac is 15 years from the onset of infection. Other series give from 14 to 19 years. Since these figures are arithmetic averages, we find of course some patients with a shorter life span and others with a longer one. Instances are recorded of patients who survived for more than 50 years after the initial infection. This is rare, however. The age at which the initial rheumatic infection occurred is, therefore, an important landmark. The chances of developing rheumatic heart disease are greatest between 8 and 11 years and decrease yearly after that. There are, however, a sufficient number who get their initial infection after the age of 11 to make the average age 17 years at onset in one adult group recently reported. It is probable, as suggested in a preceding paragraph, that the initial infection represents reinfection and that in some cases the actual onset occurred earlier than stated but was slight and escaped notice. If this is a common experience the average duration of life would probably be longer than 15 years. On the other hand, the high mortality from rheumatic heart disease in the first and second decades would probably more than compensate for such lack in the data from which this prediction was made, since they represent only patients who lived beyond the ages of childhood. The probability is that in a group comprising both children and adults the mean duration of life would be even less than 15 years. A fortunate minority (25% to 35%) of patients afflicted with a rheumatic affection have minimal cardiac lesions which heal completely. These patients probably live out the normal life span. That the disease is one of childhood and early adult life is well seen in a study of the age of death. The majority of sufferers from this disease are dead before the age of forty.

Several important events in the course help to orient one in respect to the disease as a whole. One such event is the onset of symptoms of cardiac insufficiency. These manifest themselves usually as increasing fatigue and dyspnea. Thus, tasks which formerly produced no appreciable symptoms cannot be done now without discomfort. If patients are seen at this stage of the disease, or if this date can be obtained, the probable course from this point can be charted. From the initial infection to the first clinical evidences of cardiac insufficiency the duration is, on the average, about 11 years. After this the length of life is relatively short, averaging 5 years. During this time the patient becomes more and more incapacitated and eventually heart failure ensues, unless some complication terminates the disease suddenly. Life expectancy is about three years after the first failure.

In planning the future for a young adult with rheumatic heart disease seen soon after the initial infection, it is well to remember these facts. Despite the seeming good health found in many cases it is well to advise light work and moderate exercise so that no undue strain is put upon the damaged heart. It is not wise to have a patient spend years in preparation for a trade or profession when the period of economic usefulness is relatively so short.

The natural course of rheumatic heart disease is affected materially by the rheumatic infection itself. Not only may the virulence vary from person to person so that different degrees of cardiac damage result, but in some cases the infection may be only mildly active while in others it is rapidly progressive. This factor in prognosis is extremely difficult to measure. Specific information is lacking chiefly because of the difficulty in recognizing rheumatic activity which may be present even when the patient is symptom free.

We might for the sake of convenience classify types of rheumatic heart disease, as far as the course is concerned, into the following groups:

(1) A single attack of rheumatic fever producing some cardiac damage with no subsequent recurrences. The length of life in these cases will depend on how much damage has been done to the heart. If only a moderate valve lesion is present and cardiac hypertrophy is not great, patients in this group may live for many years far beyond the average for rheumatic heart disease. For instance, mitral stenosis may be discovered during the second decade of life and the patient may still be living at 60 years. Unfortunately, most cases do not fall into this group.

(2) Recurrent attacks of rheumatic fever causing further cardiac damage with each attack. It would seem reasonable that the more attacks a patient has the shorter the life expectancy. Yet different observers have found that there is no constant relationship between the number of attacks of rheumatic fever and the time when death occurs. One possible explanation is that many of these cases would have been found to be not periodically, but continuously, active if they could have been observed more intensively. There is no sharp line dividing this group from the next.

(3) Continuous low grade activity extending over many years producing slowly progressive changes in the heart. This group and the one preceding are the usual forms in which rheumatic heart disease is seen. The course, then, in these two groups is probably as we have described it for rheumatic heart disease as a whole.

(4) Rapid fulminating course. Populations migrating into the temperate zone from the tropics and very young children are especially apt to get this particularly virulent form.

The age at onset of the rheumatic infection seems to be a factor in the subsequent duration of life. When very young children are affected the duration of life seems to be shorter. This may be explained in part by the severe fulminating forms of the disease that are seen in this age group. In adults, life expectancy decreases with advancing age in the population as a whole. From the fact that the age at onset initial infection is greater when it occurs in adult life, it follows that the subsequent duration of life would be shorter.

The duration of life seems to be unrelated to the type of valvular lesion or to the number of valves involved with the possible exception in the case of the pulmonic or tricuspid valves. When these are damaged the prognosis does not seem to be quite as favorable as with mitral stenosis alone or in combination with aortic lesions.

The frequent occurrence of complications, often fatal, should make one cautious in forecasting the course of the disease. About 6% terminate with subacute bacterial endocarditis. Life may also be shortened by intercurrent infections, particularly pneumonia. Complications directly related to the heart, such as embolism, account for about 10% of the deaths.

Auricular fibrillation is seen with greater frequency near the end of the disease. When present, therefore, it is more an indication of the advanced stage of heart disease than a serious complication in itself. Although patients with this irregularity are known to live for many years in comparative comfort, yet the average duration of life after its onset is less than 3 years.

The statements made above concerning the prognosis of rheumatic heart disease are subject to revision as more is learned of the natural course of this protracted disease. The numerical averages, particularly, should be considered as rough approximations to the truth in that they represent chiefly the experiences of one group. Also, certain variables which affect the course have been mentioned, but there are others perhaps equally important which still require study.

This brief review should indicate, therefore, the enormous difficulties that confront one in making a prognosis in a disease which extends over years and which is affected by so many variables, many of which are not thoroughly understood at the present time.

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